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Long-Term Survival of a Patient With Primary Sellar Choriocarcinoma With Pulmonary Metastases: A Case Report

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Extracranial metastasis is an unusual complication of most types of primary intracranial tumor. Approximately one-third of reported cases of primary intracranial choriocarcinoma have been associated with pulmonary tumor metastasis. The prognosis of such patients has been uniformly fatal. This report describes a probable long-term survivor of primary intracranial choriocarcinoma with pulmonary metastasis. The patient had a complete response to combination chemotherapy with cisplatin, etoposide, and bleomycin and is surviving free of disease >3 years from diagnosis.

Key words: brain neoplasm, choriocarcinoma, germ cell tumor, human chorionic gonadotropin, neoplasm metastasis, intrasellar tumor

INTRODUCTION

Choriocarcinoma constitutes a rare form of differentiated germ cell tumor arising from extraembryonic trophoblastic tissue. In combination with yolk sac tumor and embryonal carcinoma, it accounts for ~10% of germ cell tumors originating in the central nervous system (CNS) [1]. These neoplasms arise along the roof of the third ventricle, predominantly in the pineal or suprasellar regions. Extracranial metastasis from choriocarcinoma arising from the pituitary fossa has been reported previously [2–4]. This report provides further evidence of the propensity of choriocarcinoma to metastasize beyond the neuraxis.

CASE REPORT

The patient initially presented to his primary care physician at age 12 with a several month history of progressively worsening headaches. The pain was nonthrobbing in nature and was located retro-orbitally. Initial radiographic evaluation for possible sinusitis demonstrated an abnormality in the sellar region. Following the development of ptosis, anisocoria, and mild exophoria of the left eye, a lateral skull film showed enlargement of the sellar region with thinning and destruction of the bony floor. Magnetic resonance imaging (MRI) of the head demonstrated a large mixed iso- and hyper-intense (by T1 weighted signal) intrasellar lesion extending into the suprasellar cistern (Fig. 1). Cavernous sinus involvement was also noted, with encasement of the left carotid artery. The lesion did not enhance with the addition of gadolinium. No direct involvement of the optic chiasm was seen. Beta human choriogonadotropin (BHCG) and alfafetoprotein (AFP) were not obtained at the time of diagnosis.

The patient underwent a transsphenoidal resection of the sellar lesion, which demonstrated hemorrhagic necrotic tissue grossly and microscopically (i.e., nondiagnostic). The patient's third nerve paresis worsened several weeks after surgery, and he was treated with involved field radiotherapy (4,500 cGy at 180 cGy per fraction). Shortly thereafter, the patient experienced a dramatic improvement in his extraocular movements.

Nine months following surgery, the patient complained of acute chest pain. A chest radiograph demonstrated a mass and computerized tomography (CT) of the lesion showed its paramediastinal location with extension to the carina. Two smaller rightsided parenchymal lesions were also noted. A thoracotomy was performed, which demonstrated a large lesion attached to the right lower lobe in addition to two smaller lesions in the right middle lobe and right chest wall. Biopsy of the lesions was consistent with choriocarcinoma. No other primary site was found following radiographic staging of the ab-

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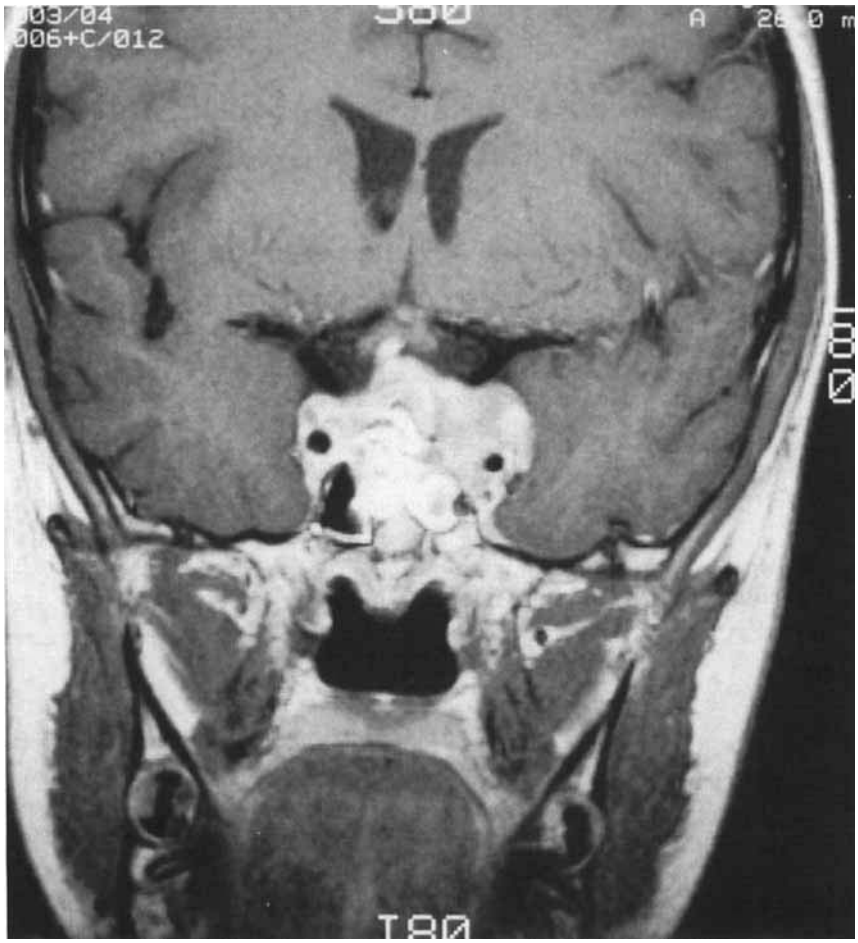

Fig. 1. Magnetic resonance image demonstrating a large mixed iso- and hyper-intense (by T1 weighted signal) intrasellar lesion extending into the suprasellar cistern.

domen and chest. A serum BHCG level obtained at this time was 58,800 IU/ml. His AFP was normal at a level of 2.5 IU/ml.

Chemotherapy consisting of four courses of cisplatin (20 mg/m^2/day for 5 consecutive days), etoposide (100 mg/m^2/day for 5 consecutive days), and bleomycin (15 mg/m^2 for 1 day) was completed, resulting in a decrease in his BHCG level to 5 IU/ml. Three months after the initiation of chemotherapy, the patient underwent second-look surgery to evaluate for residual tumor. No evidence of choriocarcinoma was found, and the patient remains in complete remission >36 months following his initial diagnosis.

DISCUSSION

Pulmonary metastasis has been reported previously in association with primary intracranial choriocarcinoma. The incidence of this complication appears to be quite high. In a recent review, of 30 patients with intracranial choriocarcinoma who underwent postmortem examinations, 13 (43%) had pulmonary metastasis [5].

Long-term survival of primary intracranial choriocarcinoma with systemic metastasis has not been previously reported. However, cisplatinum-containing regimens have been shown to be successful in the treatment of metastatic non-CNS germ cell tumors; cisplatin, etoposide, and bleomycin when used in combination achieve a 70–92% CR rate [6] and most patients with a complete response are long-term survivors. Watterson and Priest [4] reported prolonged survival of a child with CNS nongerminomatous germ cell tumor without elements of choriocarcinoma, which was metastatic. This patient was treated with chemotherapy including cis-platinum, etoposide (VP-16), vinblastine, and bleomycin. Yoshida et al. [7] recently reported a 78% response rate among 18 nongerminomatous malignant tumors of the CNS treated with cisplatin and etoposide, including two cases of cho-

riocarcinoma. Our patient was treated with a similar regimen and is, to our knowledge, the first reported case of long-term survival of metastatic intracranial choriocarcinoma.

The patient's clinical presentation with headache and evolving third cranial nerve paresis was characteristic of neoplastic lesions originating in the region of the sella tursica with cavernous sinus involvement [8]. Since the histologic specimen was nondiagnostic, a provisional clinical diagnosis of a necrotic, pituitary adenoma was made. However, when the patient developed chest masses consistent with choriocarcinoma, it became evident that the earlier intra- and suprasellar lesion was the original primary focus of disease, which involved the venous cavernous sinuses and subsequently metastasized.

Whereas mild elevation of BHCG has been noted in some patients with pure germinoma, malignant differentiated germ cell tumors may produce BHCG, alpha-fetoprotein, or a combination of the two [9,10]. High levels of either hormone usually exclude the diagnosis of pure germinoma. Obtaining serum markers at presentation of a sellar mass may thus help distinguish sellar germinomas and adenomas from malignant differentiated germ cell tumors, which would benefit from the addition of platinum-based chemotherapy.

In summary, this patient represents what appears to be the first long-term survivor of primary intracranial choriocarcinoma with pulmonary metastasis.

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